

Halpern and Copsey,³ in a comprehensive paper, recorded the varied symptomatology of the disease and the observed objective findings. The nervous, gastrointestinal and cardiovascular systems are most frequently involved and there is a tendency for objective as well as subjective findings to disappear as the acute attack subsides. Characteristics of cardiovascular involvement described are tachycardia, transient changes in electrocardiograms, and transient hypertension. Halpern and Copsey also reported three cases in which there was necrosis of renal arterioles, as well as one case of periarteritis nodosa. Attenuation of the retinal arteries occurring during attacks (associated with hypertension in some cases) has been observed.⁸ Involvement of the nervous system has been manifested by convulsions, peripheral neuritis, flaccid paralysis, amblyopias, scotomata, cranial nerve changes, respiratory paralysis and the demonstration, occasionally, of the reaction of degeneration. In addition, apathy, catatonia, sleep disturbances, delirium, psychoses and headaches have been observed to occur. Gastrointestinal tract involvement is manifested chiefly by nausea, vomiting, abdominal pain, distention, constipation, and ileus; even diarrhea and jaundice have been reported. X-ray studies during attacks reveal generalized or localized constriction or dilatation of practically any portion of the gastrointestinal tract. Fever and polymorphonuclear leukocytosis are the common systemic responses during acute attacks.

It appears probable that angiospasm could be responsible for at least a portion of the findings relative to the cardiovascular and nervous systems. Petechial hemorrhages, focal necrosis and widespread degeneration in the central nervous system have been described.^{4, 6} Restriction of the blood supply conceivably could induce changes in the motor activity of the gastrointestinal tract such as to produce clinical symptoms. Blood vessel spasm might also serve as an explanation of the infrequent observations of hematemesis and melena⁸ during the acute episodes of this disease; or, spasm of smooth muscles involving the intestinal tract directly could give rise to the gastrointestinal picture. Mason and co-workers⁵ and Nesbitt⁶ attributed findings to lesions in the autonomic nervous system. Dobriner and Rhoads² considered the muscular action of porphyrin as capable of producing the disease picture, possibly through different mechanisms since the action on the gastrointestinal tract could be inhibited by neostigmine and the capillary effect by mecholyl. Although the ability to produce spasm of smooth muscle by porphyrin compounds is well recognized,^{2, 7} clinically, a severe attack may be accompanied by a relatively low porphyrin excretion, and conversely, a severe attack may abate with continued fairly high excretion of porphyrin.³ Increased porphyrin excretion appears to be a concomitant phenomenon of this disease complex and its role as an initiating factor of the acute clinical attacks is yet to be clearly proved.

According to Hazard⁴ the effect of intravenous procaine therapy in man is the aggregate of analgesic, sympatholytic and vasodilating actions and, secondarily, of parasympathetic and anti-contracting action. In the case herein described the dramatic improvement and the duration of remission following the administration of procaine intravenously during three distinct acute clinical exacerbations of the disease suggest that the action of the procaine was something more than simple analgesia.

SUMMARY

A classical case of acute porphyria is described. Clinical and laboratory findings were characteristic of this disease complex. The patient improved dramatically with intravenous administration of procaine during three separate exacerbations.

The site of action of the procaine was not ascertained, but it is felt that its sympatholytic, anti-contracting and

secondarily parasympathetic effects were responsible for breaking down a pathologic reflex state which appears to distinguish the acute attacks.

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REFERENCES

1. Abbott, K. H., and Evans, H. S.: *Bull. Los Angeles Neurol. Soc.*, 2:20, June 1946.
2. Dobriner, K., and Rhoads, C. P.: The porphyrins in health and disease, *Physiol. Rev.*, 20:416, July 1940.
3. Halpern, R. M., and Copsey, H. G.: Acute idiopathic porphyria, *Med. Cl. N. Am.*, March 1946.
4. Hazard, R.: Present status of the physiologic actions of novocaine and its therapeutic applications, *Progress Med.*, 73:403, Dec. 10, 1945.
5. Mason, V. R., Courville, C. B., and Ziskind, E.: The porphyrins in human disease, *Medicine*, 12:355, Dec. 1933.
6. Nesbitt, S.: Acute porphyria, *J.A.M.A.*, 124:286, Jan. 29, 1944.
7. Rask, E. N., and Howell, W. N.: Photodynamic action of hematoporphyrin, *Am. J. Physiol.*, 84:363, March 1928.
8. Watson, C. J.: *A Textbook of Medicine*, 7th ed., Cecil, R. L., W. B. Saunders Co., 1947.

Bile Peritonitis Due to Spontaneous Rupture of a Dilated Intrahepatic Duct

Report of a Case

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THERE are several generally recognized ways in which bile may enter the peritoneal cavity. The more common causes of choleperitoneum are rupture of the wall of the gallbladder or extrahepatic ducts from obvious necrosis, leakage of bile following cholecystectomy, and effusion from the bile ducts in the absence of demonstrable perforation. In the case of neglected cholecystitis herewith reported, autopsy revealed an unusual location of perforation of the biliary tract. A dilated intrahepatic duct on the anterior surface of the liver ruptured spontaneously, resulting in bile peritonitis.

CASE REPORT

A white housewife, aged 66; entered the hospital on March 7, 1949, complaining of severe intermittent pain of 24 hours' duration in the right upper quadrant of the abdomen. The pain was intense, radiated to the back, right shoulder, and epigastrium, and was accompanied by frequent vomiting of white mucoid material. Two days before admission the patient had had frequency and urgency of urination and had passed several loose greenish-black stools. There was no previous history of stools or urine of unusual color. There were no chills, fever, or jaundice. The patient stated that her weight had been static and her appetite fair except for an intolerance to cabbage, beans, and fatty foods. She complained of passing considerable flatus. There had been many similar attacks, three necessitating hospitalization. In each instance the patient had been told that she had disease of the gallbladder and operation had been recommended but she refused. Cholecystography showed the gallbladder was not functioning. Previous operations included cesarean section, tonsillectomy, and four uterine curettements.

The blood pressure was 140 mm. of mercury systolic and 110 mm. diastolic, and the pulse rate was 84. The temperature was 100.4° F. Extreme tenderness of the abdomen in the right upper quadrant and epigastrium was noted, and

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there was moderate rigidity with rebound tenderness. No masses were palpable and no bowel sounds could be heard.

Results of examination of the blood and urine were within normal limits. The amylase value of the plasma was 76 units. A roentgenogram of the abdomen revealed no abnormalities and there were no opaque biliary or renal calculi.

The patient was given penicillin and general supportive therapy. The next day the condition had become much worse, and a surgical consultant advised operation. This was refused by the patient. On March 9 severe shock occurred. Some improvement followed transfusions of whole blood, but oliguria developed. Only 25 cc. of very dark urine was passed during the day. On March 10 only 8 cc. of urine was excreted, and slight jaundice was noted. The patient was semicomatose throughout the day and she died soon after midnight.

The significant autopsy findings pertaining to the liver and gallbladder were as follows: The peritoneal cavity contained approximately 800 cc. of cloudy, yellow, foul-smelling fluid. There were no adhesions. The gallbladder was greatly dilated and tense. There was pronounced dilatation of the cystic, hepatic and common ducts, the latter measuring 2 cm. in diameter. The gallbladder and all extrahepatic bile ducts were filled with numerous faceted gallstones. One calculus, 2 mm. in diameter, was lodged in the ampulla of

Vater and completely occluded it. The mucosa of the gallbladder and bile ducts was intact and showed no evidence of perforation. A thick, bile-stained, fibrinous exudate covered the anterolateral surface of the right lobe of the liver and extended over the anterior surface of the gallbladder. The undersurfaces of this lobe and the gallbladder were entirely free of exudate, as was the rest of the liver to the left of the falciform ligament. In one area the exudate seemed somewhat thicker than elsewhere. In a section cut through the area, there appeared to be a greatly dilated intrahepatic duct lying just beneath the surface. It was 6 mm. in diameter, and the distal end was covered only with the fibrinous exudate noted on the surface of the liver. All intrahepatic ducts were greatly dilated and were filled with cloudy green bile.

Two sections of liver were studied. These were taken to include the dilated intrahepatic bile duct noted near the anterior surface. Along the lining surface of this space there was a thick layer of fibrous, partly hyalinized connective tissue. There was no epithelium lining the space. The liver tissue surrounding the cystic space showed varying degrees of degenerative change. The liver sinusoids were dilated and many were filled with blood. The bile capillaries were moderately dilated and a few contained inspissated bile.

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